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Case Report

Typical presentation of pyoderma gangrenosum on an atypical site

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ABSTRACT

Pyoderma gangrenosum (PG) is an uncommon inflammatory skin condition that presents with rapidly progressive, painful ulcers with undermined borders. While PG commonly affects the lower extremities, it may rarely appear at atypical sites, posing diagnostic challenges. Herein, we report the case of a 45-year-old, healthy male who presented with a solitary, rapidly expanding ulcer over the anterior chest wall. Initially misdiagnosed as a cutaneous infection, further history-taking and investigations revealed features consistent with PG. The patient responded well to topical wound care and systemic corticosteroids, with complete resolution of the ulcer after a seven-week period. This case highlights the importance of considering PG in the differential diagnosis of ulcers over the anterior chest wall, especially when standard therapies for infections fail to yield improvement.

Key words: pyoderma gangrenosum; chest wall ulcer; neutrophilic dermatoses

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare, idiopathic, recurrent, ulcerative, non-infective chronic skin disorder with an infiltrate that is predominantly neutrophilic. It has unique morphologic features and is often associated with systemic diseases [1]. Its etiology is unknown, and its pathogenesis is highly uncertain. An underlying immunological abnormality is currently favored because of the frequent association of PG with systemic diseases. Although the most common site is the lower extremities, other areas may be involved. The development of an isolated lesion of PG over the chest region is very rarely reported [2-4]. We report, herein, one such case of PG involving the chest wall.

CASE REPORT

A 45-year-old male presented with a rapidly progressing, painful ulcer over the left side of the chest lasting for the previous two weeks. It was associated with bleeding and without pus discharge. There was no history of trauma, fever, or associated systemic diseases. Cutaneous examination showed an ulcer measuring 22 x 14 cm with undermined edges, violaceous irregular margins, and red granulation tissue on the floor, located over the left side of the anterior chest wall (Fig. 1). He had received several courses of antibiotics without improvement. The differentials of pyoderma gangrenosum, vasculitis, and necrotizing fasciitis were considered. Blood investigations showed an elevation of inflammatory markers (ESR and CRP). His chest x-ray was normal. Fungal and bacterial cultures showed no growth. The pathergy test was negative. A biopsy from the edge of the ulcer showed dermal neutrophilia and mixed inflammatory infiltrates, which favored the diagnosis of PG (Fig. 2). The patient was started on prednisolone 60 mg for two weeks, followed by 50 mg for the next two weeks, along with daily dressing with a chlorhexidine gauze dressing. During follow-up after a month, the patient showed 60% improvement in the lesion. The dose of prednisolone was tapered to 40 mg for three weeks. During the next follow-up, the lesion

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Figure 1: An ulcer with undermined edges, a violaceous border, and granulation tissue seen on the anterior chest wall.

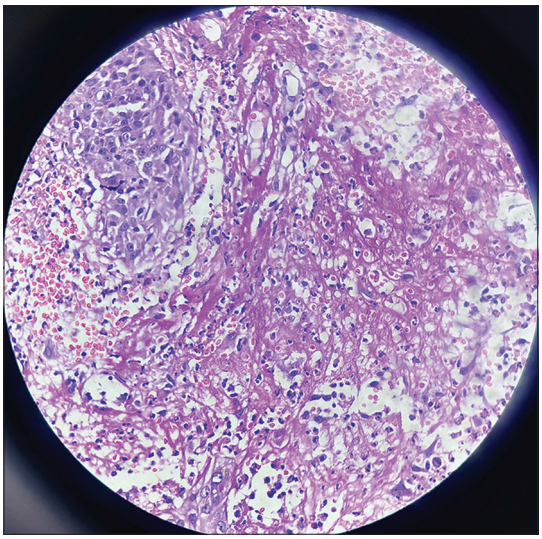

Figure 2: Massive dermal neutrophilic infiltration mixed with lymphocytic inflammatory infiltrate (H&E; 40x).

almost completely healed, with some hypertrophic scars (Fig. 3).

DISCUSSION

PG is a type of neutrophilic dermatosis that classically presents as an ulcer, but there are other variants seen, such as the bullous, vegetative and pustular types.

Figure 3: Hypertrophic scars formed after seven weeks of treatment

The ulcers are extremely painful due to necrosis of the skin and subcutaneous tissues. The ulcers heal by re-epithelialization from the edges and, ultimately, leave behind a cribriform atrophic scar.

In 80% of cases, PG involves the extremities, and around 11% involves the trunk [5], and on the trunk, the involvement of the chest wall alone is very rare. Herein, we report a case of PG with a typical presentation at an atypical site. The disease is idiopathic in about 25–50% of cases [6], whereas around 50% cases are associated with systemic diseases such as inflammatory bowel disease, rheumatoid arthritis, hematological malignancies, and HIV. These associated conditions can occur before, after, or simultaneously [7].

The pathergy phenomenon is a localized, misdirected effector host cell response to trauma, which induces cutaneous tissue changes in an individual with a compromised immune response. Pathergy test is often positive in cases of PG associated with systemic illnesses [8]. In our case, however, the pathergy test was negative, which we believe was because there was no underlying systemic disease.

Su et al.'s diagnostic criteria for PG should meet two major and two minor requirements [9]. The major criteria are 1) a painful necrolytic cutaneous ulcer with a violaceous, undermined and irregular border that progresses rapidly; 2) other causes of ulcerations to be excluded. The minor criteria are 1) a pathergy phenomenon history or presence of cribriform scarring; 2) systemic illnesses associated with pyoderma gangrenosum; 3) histopathology showing (lymphocytic vasculitis ± mixed inflammation ± sterile dermal neutrophilia); and 4) a response to treatment (quick

response to systemic corticosteroid therapy). Our patient fulfilled both major and two of the minor criteria, namely, the histopathologic findings and rapid response to systemic corticosteroids.

The treatment modalities include a combination of systemic medications, topicals, and wound care. It depends on the extent of lesions and associated systemic diseases. In all cases, wound care in the form of wet compresses and low-pressure dressings is mandatory. For limited superficial disease, topical management would be enough. Topical calcineurin inhibitors (such as tacrolimus) or corticosteroids may be administered directly to the afflicted skin.

For extensive disease and limited disease not responding adequately to wound care and topicals, therapy with systemic corticosteroids should be started. In addition, systemic immunosuppressants such as cyclosporine, azathioprine, or mycophenolate mofetil may be added. For severe or refractory cases of PG, biologic agents such as adalimumab, infliximab, or etanercept can be considered.

CONCLUSION

This case report highlights a rare presentation of PG involving the anterior chest wall, which is often challenging to diagnose due to its resemblance to other ulcerative conditions. Through a proper clinical examination, histopathological analysis, and the exclusion of other potential causes, the diagnosis was confirmed. Prompt initiation of systemic corticosteroid therapy combined with wound care management led to significant improvement in the patient's condition, emphasizing the importance of early intervention in managing PG.

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Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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